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Causes of Late Deaths After Pediatric Cardiac Surgery

A Population-Based Study

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Objectives We examined the causes and modes of late death after pediatric cardiac surgery.

Background The late mortality of patients operated on for congenital heart defect (CHD) is comprehensively unexamined. In this study, the causes of death were examined to obtain further knowledge of the morbidity of the patients.

Methods We studied all late deaths of patients operated on for CHD in Finland during the years 1953 to 1989. We calculated the survival of patients, identified the causes of deaths from death certificates, and examined the modes of CHD-related deaths. We compared the survival and the causes of non–CHD-related deaths to those of the general population.

Results Of the 6,024 patients who survived their first operation, 592 (9%) died during the 45-year follow-up period. The progress of treatment was seen in the survival of the patients operated on in different decades. The cause of death was confirmed with postmortem examination in 474 (81%) cases. The majority of patients (397, 67%) died owing to the CHD. Furthermore, non–CHD-related mortality was twice as high (risk ratio 1.9, 95% confidence interval 1.5 to 2.4) as expected. The main mode for CHD-related death was heart failure (40%). Other modes included perioperative (26%), sudden (22%), and cardiovascular (12%) deaths. The number of deaths caused by neurological and respiratory diseases was higher and the number of accidental deaths was lower than expected.

Conclusions The survival of patients was lower than that of the general population (relative 45-year survival 89%). Most patients died owing to CHDs, but non–CHD-related mortality was also high. (J Am Coll Cardiol 2007;50:1263–71)

This present study is the first population-based evaluation of the causes of late death after pediatric cardiac surgery. The comprehensive coverage and reliability of the Finnish population register and the Finnish research register of pediatric cardiac surgery enabled us to complete this unique study. Our study includes all children operated on for congenital heart defects (CHDs) in Finland during the years 1953 to 1989, and we were able to confirm the late outcome of 96% of them.

For the first time, the causes of death of CHD patients could be compared with those of the general population. Previously, the causes of late deaths were reported incompletely only as part of follow-up investigations (1–5).

The results of pediatric cardiac surgery are good, but mortality still remains higher than in the general population (6,7). Most of the patients die owing to their cardiac defect. With active and well-timed interventions, however, the lifespan of these patients can be prolonged. Therefore, an understanding of the causes and the mechanisms of death play an important role in improving late prognosis.

The gender difference in the late mortality of CHD patients has not yet been studied. Gender could be a factor in the prognosis of such patients, because significant differences have been found in the mechanisms of arrhythmias between genders (8), and sudden death due to coronary heart disease is more common among male patients (9).

Methods

According to the Finnish Research Registry of Pediatric Cardiac Surgery, a total of 7,240 cardiac operations were performed on 6,460 children in Finland during the years 1953 to 1989.

The current state of patients on the closing day of this study (October 28, 1998) and the dates of death and emigration were obtained from the Finnish Population Registry Centre. The causes of late death were identified from death certificates obtained from Statistics Finland. If something unclear ap-

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Abbreviations and Acronyms

ASD = atrial septal defect
CHD = congenital heart defect
CI = confidence interval
CoA = coarctation of aorta
PDA = patent ductus arteriosus
PS = pulmonary stenosis
RR = relative risk
SMR = standardized mortality ratio
TGA = transposition of the great arteries
TOF = tetralogy of Fallot
UVH = univentricular heart
VSD = ventricular septal defect

peared in the death certificate, we examined the patient's medical records.

The patients were divided into different heart defect groups according to a previously published hierarchy based on their primary diagnosis (6). We investigated separately the mortality of patients with patent ductus arteriosus (PDA), coarctation of aorta (CoA), atrial septal defect (ASD), ventricular septal defect (VSD), tetralogy of Fallot (TOF), transposition of the great arteries (TGA), and univentricular heart (UVH) who had survived their first operation (i.e., were alive >30 days after).

The deaths were first divided by their main cause into CHD-related and non–CHD-related deaths; CHD-related causes included deaths caused by diseases with ICD-10 (International Classification of Diseases) diagnosis numbers Q20 to Q28 (in old cases ICD-9 numbers 745 to 747). All other causes were classified as non–CHD-related.

An existing classification of the cardiac cause of deaths (10) served to divide the CHD-related deaths into 4 groups: heart failure, sudden, perioperative, and cardiovascular death. Sudden death was defined as death due to cardiovascular causes within 1 h of onset (or significant worsening) of symptoms or unwitnessed death during sleep (11). The perioperative deaths included all early postoperative (within 30 days) deaths due to patient's second, third or fourth operation. The cardiovascular group included all CHD-related deaths that could not be classified into the other 3 groups.

This study was conducted with the permission of the Finnish Ministry of Social Affairs and Health.

Statistical methods. Time-related mortality was assessed with survival analysis. Survival was calculated for all patients and separately for gender and diagnostic groups. The survival rate of patients operated on in different decades (1950s to 1980s) was also calculated separately to evaluate the progress of treatment. We compared the survival rates of different gender and decade groups and analyzed survival with and without non–CHD-related causes of death. The survival statistics were calculated with SURV3, the latest version of the survival analysis package developed at the Finnish Cancer Registry (12).

We compared each patient's individual survival with that of the age-, gender-, and time-matched general population. For every patient and for every year of follow-up, we obtained the mortality of the general population. The expected probability of surviving 1 year was obtained for all individuals who, regardless of their survival status, were not censored before the beginning of the interval. We used the Hakulinen method (13) for combining the individual information with an estimate for the expected survival of the group.

The relative risk (RR) (with 95% confidence intervals [CIs]) analysis was used in evaluating the differences between observed versus expected numbers of deaths and between genders. If the CI of RR did not include 1.0, the difference was considered significant. Furthermore, the narrower the range of interval, the more significant was the difference.

The non–CHD-related causes of death of the patients were compared with the cause-specific mortality of the general population. The results appear as numbers of deaths observed and expected and as standardized mortality ratios (SMRs). The SMR expresses the ratio of observed causes of death to that expected on the basis of overall mortality rates in the general Finnish population. The expected numbers of deaths were standardized for age, gender, and time. The Poisson distribution was used to calculate the 95% CI. The significance of SMR was calculated with the approximation of Byar for the exact Poisson test (14).

Results

On the closing day of the study, the condition of all 6,460 operated patients was confirmed from the population registry: 5,193 (80%) patients were known to be alive and living in Finland. Altogether, 1,028 (16%) patients had died; 436 (7%) died early (≤30 days), and 592 (9%) died late (>30 days) after the first operation. The outcome of 239 (4%) surviving patients remained unknown, because they were either unidentifiable from the population registry or had emigrated. However, 134 emigrated patients were included in the survival analysis, because their emigration date was known and served as the censoring date. Thus, the survival analysis included altogether 5,919 patients (98% of the 6,024 patients), all who were alive, had emigrated, or died late after surgery.

Of these 5,919 patients, 2,732 were male and 3,187 (54%) female. The CHDs were distributed unequally between genders. The vast majority (71%, 1,377 of 1,932) of the PDA patients and 60% (461 of 767) of ASD patients were female. Male patients more often exhibited CoA (66%, 583 of 880), TOF (60%, 246 of 413), and TGA (66%, 172 of 259). The proportion of male and female patients with VSD and UVH were equal.

A total of 592 patients died late after surgery. The mortality was highest shortly after the first operation. One hundred sixty-four (28%) deaths occurred during the first year, and almost one-half of them (n = 79, 48%) occurred during the first 3 months after surgery. The patients that died were significantly younger at the time of their first operation than the patients that survived, 3.9 (median 1.3, range 0 to 15) years versus 5.4 (median 5.1, range 0 to 15) years ($p < 0.001$), respectively. The mean age of late death

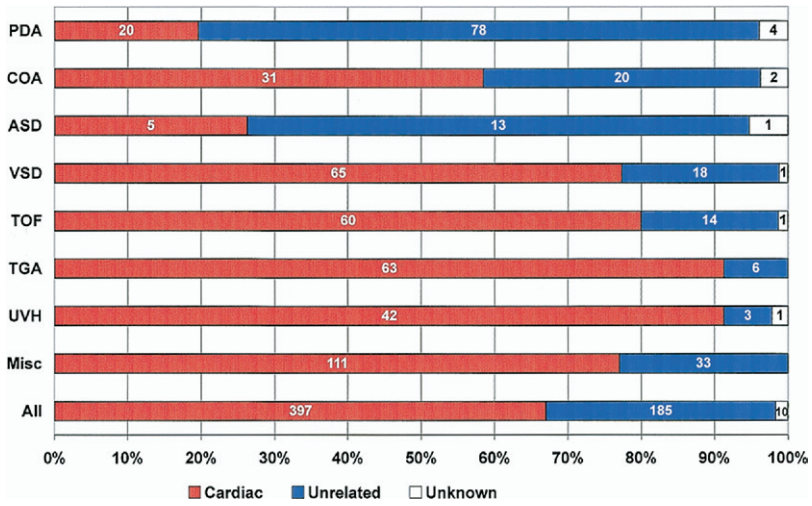

Figure 1 **Proportion of Causes of Death**

Numbers in blocks define the actual number of deaths in each diagnostic group. ASD = atrial septal defect; CoA = coarctation of aorta; PDA = patent ductus arteriosus; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; UVH = univentricular heart; VSD = ventricular septal defect.

was 13.3 (median 10.2, range 0.1 to 53) years. The patients died, on average, 9.4 (median 5.8, range 0.08 to 39) years after their first operation.

The cause of death was identified in 582 patients and had been confirmed with postmortem examination in 474 (81%) cases. A total of 397 (67%) deaths were related and 185 (31%) were unrelated to CHDs. The cause of death was undetermined in 10 (2%) patients. In most diagnostic groups, CHD-related causes of death dominated. Only patients with PDA and ASD died more often owing to non–CHD-related causes (Fig. 1).

The survival of the patients was lower than that of the general population (Fig. 2). The relative survival for 45 years of patients was 89% (in relative survival, the survival of the general population is always 100%). When only CHD-related deaths were included in the analysis, 45-year survival was 96%. Most patients died owing to their heart defects, but the number of non–CHD-related deaths was also higher than expected: 185 versus 97, respectively. The RR for non–CHD-related death was 1.9 (95% CI 1.5 to 2.4).

The overall relative survival of male patients was lower than that of female patients ($p = 0.042$) (Fig. 2).

When only CHD-related causes of death were included in the analysis the difference in survival disappeared. Within the diagnostic groups, we found no significant differences in survival between genders either in overall mortality or CHD-related mortality.

The survival rates of all patients operated on decreased decade after decade. This reflects the fact that more children with severe CHDs were treated during the last decades than in the earlier decades of our study period. During the 1960s, 1,347 patients were operated on, and most of them exhibited simpler defects than did the 2,418 patients operated on during the 1980s (Fig. 3).

The survival of those patients with PDA who underwent surgery in the 1980s was significantly poorer than that during earlier decades ($p < 0.001$). This result was due to the difference in the number of neonates operated on: 29% (186 of 631) in the 1980s, and only 4% (22 of 525) in the 1970s. The difference in survival rates between decades in PDA patients and also in all patients disappeared when non–CHD-related causes were excluded, indicating that

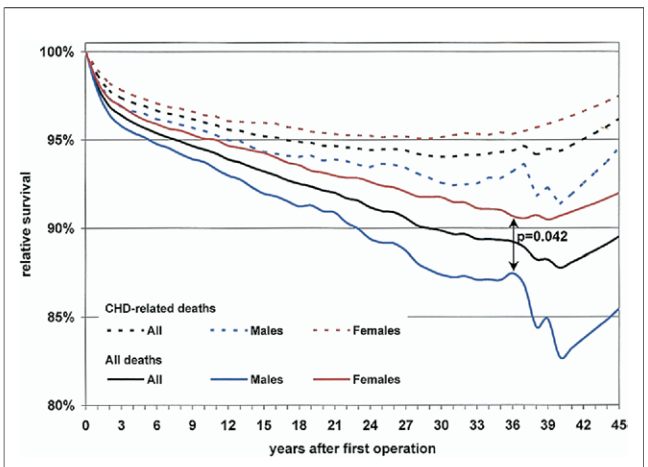

Figure 2 **Relative Survival (Survival of Patients/ Survival of General Population) of All Patients**

Survival of general population is 100%. **Dotted lines** represent survival for congenital heart defect (CHD)-related causes. Follow-up started from the patient's first operation.

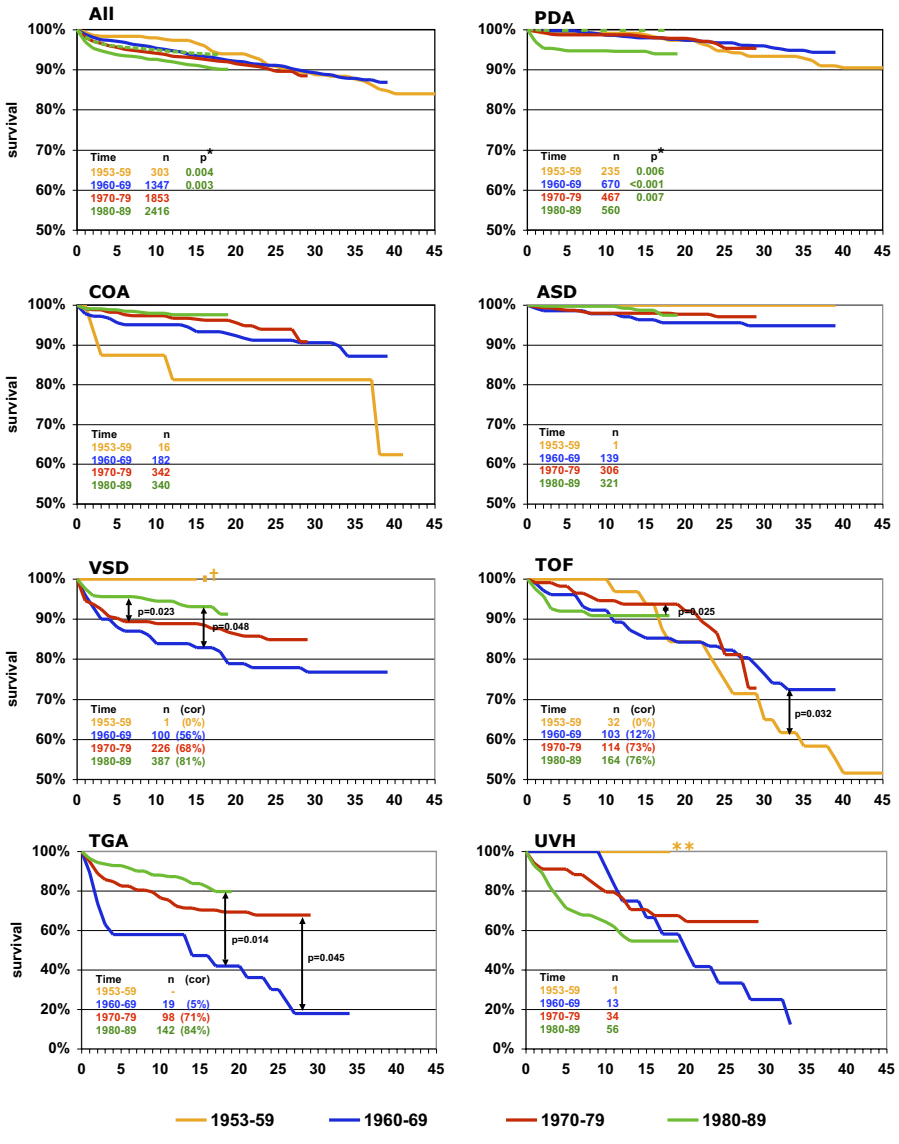

Figure 3 **Survival of Patients After Their First Operation**

Division is by the decade during which the patient underwent the first surgery. The **dotted green line** in the figures for all and for PDA patients represents the survival of the 1980s, when only CHD-related deaths were included in the analysis. Follow-up started from the patient's first operation. In VSD, TOF, and TGA figures, the percentages marked as **"cor"** represent the proportion of patients whose first operation has been corrective. *p = significance of difference when compared with 1980s. †Patient died 15 years after closure of PDA; VSD was not surgically corrected. **Patient emigrated. Abbreviations as in Figures 1 and 2.

most of the neonates were small, premature babies dying from non-cardiac causes (Fig. 3).

As expected the relative survivals of patients with CoA, ASD, VSD, and TGA improved over the years (Fig. 3).

The survival of the patients with TOF who underwent surgery in the 1980s was worse than of those in the 1970s. The increase in mortality resulted mainly because patients were younger at the time of their operation, mean 2.8 versus 6.3 years ($p < 0.001$), respectively. Also within the patients operated on during the 1980s the difference in age was significant. The mean age at operation for survivors was 3.0 and 1.1 for those who died late after surgery, $p = 0.015$.

The survival of the patients with univentricular heart was poor. The 19-year survival for those operated on in the 1970s was better than for those operated on in the 1980s, 68% versus 55%, respectively. The higher mortality in the 1980s was explained by the fact that more and more complicated defects came within the reach of surgery. For instance, during the 1980s, of the 56 operated UVH patients 8 (14%) had a combination UVH, TGA, and CoA, whereas in the 1970s only 1 (3%) of the 33 operated UVH patients had this complex defect.

The survival curves of different diagnostic groups and decades appear in Figure 3.

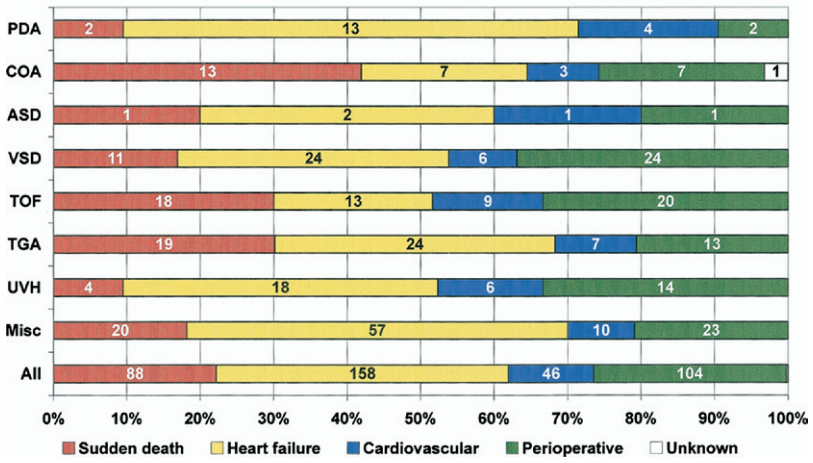

Figure 4 Proportion of Modes of CHD-Related Death in Different Defect Groups

Perioperative = death within 30 days after second, third, or fourth operation. **Numbers in blocks** define the actual number of deaths in each group. Abbreviations as in Figures 1 and 2.

Mode of CHD-related deaths. Most CHD-related deaths resulted from heart failure (158 of 397, 40%) (Table 1), which was the main mechanism of death in PDA, TGA, and UVH groups (Fig. 4). One-fourth (38 of 158) of the patients who died owing to heart failure had been previously diagnosed with pulmonary hypertension.

A total of 104 (26%) late CHD-related deaths resulted from a second, third, or fourth heart operation. The majority of perioperative deaths (54, 52%) occurred early (within 30 days) after the corrective operation among patients whose first operation had been palliative; 31 patients died early after re-operation, and 19 died after palliative surgery.

Surgery was the main mode of death among patients with TOF (20 of 60 deaths); 11 died early after the corrective operation, 7 after re-operation, and 2 after palliative operation.

Of the CHD-related deaths, 88 (22%) occurred suddenly. The mean age at sudden death was 19.3 years, whereas it ranged from 8.2 to 12.8 years in the other groups (Table 1). The vast majority of these patients had undergone corrective surgery (60 of 88, 68%). In most cases, sudden death resulted from arrhythmia, because no other reason for death could be found in postmortem examination (84%, 74 of 88 were examined). The majority of these patients had arrhythmias in their medical history. The other underlying causes of sudden death appear in Table 2.

Sudden death was the most common mode of death among patients with CoA (13 of 31, 42%) (Fig. 4). Of the 13 CoA patients who died suddenly, 5 (38%) exhibited untreated valvular aortic stenosis (AS). In 4 cases, the underlying cause was a ruptured or dissected aorta.

Among patients with TOF (18 of 60) and TGA (19 of 63), 30% of CHD-related deaths were sudden; 6% (15 of 246) of male patients with TOF and 10% (17 of 171) of male patients with TGA died suddenly. The proportion of sudden deaths among female patients was 2% in both groups (3 of 168, 2 of 171, respectively). The RR of sudden death for male patients was 3.4 (95% CI 1.004 to 11.6) in the TOF group and 4.3 (95% CI 1.02 to 18.3) in the TGA group. When these 2 diagnostic groups were combined, the RR of sudden death for male patients was 3.9 (95% CI 1.5 to 9.9).

The only death caused by anti-coagulation therapy was also sudden: 1 patient with an operated CoA and a prosthetic aortic valve died owing to subarcachnoidal bleed-

Table 1 Mode of Cardiac Death

	Deaths		Median Age at 1st Op (yrs) (range)	Median Follow-Up Time (yrs) (range)	Median Age at Death (yrs) (range)
Heart failure	158	40%	0.7 (0–14.9)	2.1 (0.1–38.9)	5.2 (0.1–52.1)
Perioperative*	104	26%	0.4 (0–14.4)	2.7 (0.1–30.9)	3.9 (0.1–41.3)
Sudden death	88	22%	5.0 (0–14.3)	12.7 (0.2–37.0)	18.4 (0.2–47.6)
Cardiovascular	46	12%	2.3 (0–14.9)	2.0 (0.1–34.2)	7.6 (0.1–45.3)
Unknown	1	0%	12.1	1.1	13.2
All	397	100%	0.9 (0–14.9)	4.3 (0.1–38.9)	7.7 (0.1–52.1)

*Death within 30 days after second, third, and fourth operation (Op).

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Table 2 Causes of Sudden and Cardiovascular Deaths

	n
Cause of sudden death	
Arrhythmia/heart failure/unknown	73
Rupture of aorta	5
Pulmonary emboli	3
Myocardial infarct	2
Shunt occlusion	2
Rupture of an aneurysm of MPA	1
Subarchacnoidal bleeding	1
Pulmonary hemorrhage	1
All	88
Cardiovascular causes of death	
Stroke	11
Arrhythmia	6
Pulmonary emboli	6
Endocarditis	4
Brain abscess	4
Thrombosis	4
Myocardial infarct	3
Rupture of an aneurysm of PDA	2
Valve prothesis complication (aortic)	2
Pulmonary hemorrhage	2
Pericarditis	1
Myocarditis	1
All	46

MPA = main pulmonary artery; PDA = patent ductus arteriosus.

ing. The patient's treatment with warfarin was complicated by alcohol abuse.

Cardiovascular diseases related to congenital defects (Table 2) caused 46 (12%) deaths. In this subgroup, 3 diseases dominated: stroke (11 deaths), arrhythmia (6), and pulmonary emboli (6). Endocarditis caused 4 deaths, accounting for 1% of all CHD-related deaths. The overall incidence of lethal endocarditis was 0.07% (4 of 6,024). Two of these 4 patients had a bicuspic aortic valve, 1 exhibited re-coarctation, and 1 patient with UVH failed to recover from the Fontan operation. Brain abscess caused 4 deaths, 2 of which occurred among patients with pulmonary hypertension, and the other 2 patients had non-corrected cyanotic heart defects (TGA and TOF). Three patients died owing to myocardial infarct: 1 suffered from severe familial hypercholesterolemia (AS), 1 had anomalous coronary arteries (VSD+CoA), and 1 patient (CoA, AS, and Turner's syndrome) had an occluded coronary artery. In 2 cases, the myocardial infarction was classified as non–CHD-related: the first involved a 44-year-old patient with operated CoA, severe diabetes with several complications, and a kidney transplant; and the second was a 36-year-old patient with operated PDA. Pulmonary hemorrhage was the cause of death of 2 patients, both of whom had Eisenmenger's syndrome. In addition, 1 patient with Eisenmenger's syndrome died suddenly due to massive acute pulmonary bleeding.

Causes of non–CHD-related deaths. The cause of death in 185 patients was unrelated to cardiac malformation. Diseases caused 127 deaths, and 58 deaths were violent. We found no differences between genders in the causes of non-CHD deaths.

The patients died of respiratory and neurological diseases significantly more often than did the general population (Table 3). Of the 17 respiratory deaths, 11 resulted from pneumonia in mentally retarded patients (6) and infants (5). The most common neurological causes of death were epilepsy (6) and meningitis (4).

The patients died in accidents less often than the general population. The number of accidental deaths was 28 in the patient population; the expected value was 44 (SMR 0.64, 95% CI 0.42 to 0.92).

The number of suicides was similar to the number expected. All 23 patients who committed suicide had undergone corrective surgery. The patients had undergone surgery at least 5 (mean 16.8) years before death (9 PDA, 3 CoA, 3 pulmonary stenosis [PS], 4 VSD, 3 ASD, 1 TOF). Two suicides were directly connected to heart defects: 1 ASD patient exhibited severe arrhythmias resistant to treatment, and 1 patient with PS took her own life rather than agree to another operation.

Discussion

In the present population-based study, we examined all late deaths after pediatric cardiac surgery in Finland. The accurate research registry of pediatric cardiac surgery in Finland guarantees the reliability and coverage of the study. The number of patients lost to follow-up was 239, only 4% of all 6,024 who survived their first cardiac operation.

Statistics Finland helped us identify the cause of death in 98% of cases. Of the causes, 81% were confirmed with postmortem examination, considered a valuable way of investigating the mechanism of death even today (15).

The number of deaths was high throughout the follow-up years when compared with general population. The mortality was highest within the first year of follow-up. The high mortality during the first years appears in all previously published survival curves (2,5,7). The survival of male patients was poorer than that of female patients. As far as we know, this gender difference has not been previously published. The obvious cause was the difference in the seriousness of the defects.

In most diagnostic groups, the long-term results improved with every decade. However, in some groups the survival was worse in patients operated on in the 1980s. The lower survival rate was caused by more complicated defects and younger age at the time of surgery, both of which are known risk factors for mortality (16). The early mortality after procedures made for neonates has decreased in 1990s and 21st century (17). Yet only the future will show whether the improved early survival of infants in the current era will also lead to decreased late mortality.

Table 3 Observed and Expected Causes of Deaths Unrelated to Congenital Heart Defect

	All Deaths (n = 185)					Age at Death <20 yrs (n = 110)					Age at Death >20 yrs (n = 75)				
	Patients	Expected	SMR	95% CI	p Value	Patients	Expected	SMR	95% CI	p Value	Patients	Expected	SMR	95% CI	p Value
Diseases	127	110.85	1.15	(0.96–1.15)		87	75.93	1.15	(0.92–1.41)		40	34.92	1.15	(0.82–1.56)	
Infectious diseases	1	2.84	0.35	(0.01–1.96)		1	2.18	0.46	(0.01–2.56)		0	0.66	0.00	(0.00–5.59)	
Neoplasms	18	19.30	0.93	(0.55–1.47)		7	8.67	0.81	(0.32–1.66)		11	10.64	1.03	(0.52–1.85)	
Endocrine, nutritional, and metabolic diseases	5	3.62	1.38	(0.45–3.22)		1	1.96	0.51	(0.01–2.84)		4	1.67	2.40	(0.65–6.13)	
Diseases of the nervous system	14	6.26	2.24	(1.22–3.75)	0.01	9	3.99	2.26	(1.03–4.28)	0.04	5	2.27	2.20	(0.72–5.14)	
Diseases of the circulatory system	11	11.89	0.92	(0.46–1.66)		3	2.26	1.33	(0.27–3.88)		8	9.63	0.83	(0.36–1.64)	
Diseases of the respiratory system	17	4.59	3.70	(2.16–5.93)	<0.001	14	3.00	4.67	(2.55–7.83)	<0.001	3	1.60	1.88	(0.39–5.48)	
Diseases of the digestive system	3	2.09	1.44	(0.30–4.19)		2	1.09	1.83	(0.22–6.63)		1	0.99	1.01	(0.03–5.63)	
Diseases of the genitourinary system	1	1.10	0.91	(0.02–5.07)		1	0.62	1.60	(0.04–8.99)		0	0.47	0.00	(0.00–7.85)	
Congenital malformations and chromosomal abnormalities	22	23.08	0.95	(0.60–1.44)		20	21.76	0.92	(0.56–1.42)		2	1.32	1.52	(0.18–5.47)	
Other diseases	27	29.74	0.91	(0.60–1.32)		25	28.44	0.88	(0.57–1.30)		2	1.29	1.55	(0.19–5.60)	
Unknown	5	2.57	1.94	(0.63–4.54)		4	1.81	2.21	(0.60–5.66)		1	0.76	1.31	(0.03–7.33)	
Alcohol–related diseases	3	3.77	0.80	(0.16–2.33)		0	0.15	0.00	(0.00–24.59)		3	3.62	0.83	(0.17–2.42)	
Accidents and violence	58	74.14	0.78	(0.59–1.01)		23	34.07	0.68	(0.43–1.01)		35	40.08	0.87	(0.61–1.21)	
Accidents	28	44.04	0.64	(0.42–0.92)	0.01	16	25.84	0.62	(0.35–1.01)		12	18.20	0.66	(0.34–1.15)	
Suicides	23	22.51	0.98	(0.62–1.47)		5	5.48	0.91	(0.30–2.13)		18	18.03	1.00	(0.59–1.58)	
Homicides	4	4.08	0.98	(0.27–2.51)		2	1.74	1.15	(0.14–4.15)		2	2.34	0.82	(0.10–3.09)	
Unknown	3	2.31	1.30	(0.27–3.80)		0	0.90	0.00	(0.00–4.10)		3	1.41	2.12	(0.44–6.22)	
Other	0	0.20	0.00	(0.00–18.44)		0	0.11	0.00	(0.00–33.54)		0	0.09	0.00	(0.00–40.99)	

CI = confidence interval; SMR = standardized mortality ratio with 95% confidence limits.

The most common cause of death among patients was the heart defect. Only patients with ASD and PDA died more often owing to non–CHD-related causes. The mortality of ASD patients resembled that of the general population. Our study confirmed the previously reported higher-than-expected mortality of the PDA patients (6,7), which resulted from non-cardiac diseases. Thus, the ASD and PDA patients who underwent surgery could be considered cured.

Mode of CHD-related deaths. The most common mode of CHD-related death was heart failure. Most of the patients with lethal heart failure had diagnosed pulmonary hypertension. Part of the late mortality could perhaps have been avoided with more careful follow-up and subsequent interventions, thus preventing the further rise of pulmonary resistance.

In our subjects, the number of late deaths due to surgery was high and resulted mainly from the patient's subsequent corrective operation, whereas the follow-up began from the first—often palliative—operation (e.g., shunt in TOF).

Sudden death occurred most often in older patients with corrected heart defects and with known high risk of sudden death (18). The overall survival of patients with operated CoA was good: over 90% for 30 years. Yet the risk of sudden death was high. This obligates health professionals to ensure the careful follow-up and treatment of patients; the survivors of CoA repair are fixed, not cured (19).

Surprisingly, we discovered that male patients with corrected TOF and TGA died suddenly more often than female patients. The number of sudden deaths was small, and the significance of the difference was weak, but it existed nevertheless. When the 2 diagnostic groups were combined, the significance of the difference strengthened. Gender might be one of the fatal factors that contribute to sudden death in CHDs. Yet the causes of the difference need to be examined. This same kind of gender differences in sudden death has been published among adults with different heart diseases. For instance, male patients with coronary heart disease and male athletes die suddenly more often than female patients (9,20). In addition, the types of arrhythmias differ between men and women (8).

In the cardiovascular mode of CHD-related death, some data aroused our attention. The overall incidence of lethal endocarditis was only 0.07%. None of the deaths due to endocarditis occurred in patients with a valve prosthesis, although they are believed to carry a high risk for it (21). Yet, the other 2 patient groups at high risk for infective endocarditis were among our subjects: patients with aortic valve disease and cyanotic heart defect (21).

The number of deaths due to bleeding after anticoagulation therapy was very low in our study, confirming the previously reported low incidence of fatal bleeding among young patients (22). When only mortality for endocarditis and bleeding is concerned, the artificial heart valve seems a safe alternative for young patients.

Previous studies have reported that patients with CoA are at high risk for death due to coronary disease and cerebrovascular accidents (1). Our findings, however, differed. Among the patients who underwent a successful operation for CoA (n = 887), only 1 death resulted from coronary heart disease. Even this death was classified as non–CHD-related, because of the patient's severe diabetes and kidney transplant, which have been reported to increase one's risk for all cardiovascular diseases (23). Moreover, only 1 of the patients died owing to a cerebrovascular accident; this death resulted from the combination of warfarin and alcohol. In earlier reports, a younger age at CoA repair has protected patients against lethal complications (1). Our study confirmed this, because all of our patients underwent surgery before their 15th birthday.

Unrelated deaths. The greatest difference between the patients and the general population appeared in the deaths caused by respiratory diseases. Most patients within this group died owing to pneumonia. High mortality due to pneumonia has often been connected to mental retardation (24) and seriously ill infants, as was the case in our study.

Neurological diseases caused a relatively high number of deaths. High mortality due to neurological causes partly resulted from other malformations and genetic syndromes with neurological disorders, which are quite common among patients with CHD. Some of the patients, however, suffered from brain damage caused by the heart defect itself or by cardiac surgery. Especially in the earlier days of cardiac surgery, cardiopulmonary bypass and related techniques, although enabling the repair of the heart itself, were not so gentle as to avoid potential brain damage entirely.

The number of accidental deaths was low. This might have resulted from the influence of parental over-protection, usually reported to cause only negative effects (25). One might also interpret this phenomenon as a reflection of the physical inability of the patients to engage in risky hobbies or hard physical work that could lead to accidental death. Alternatively, patients with a surgically corrected heart defect might appreciate life enough to avoid extreme hobbies and careless driving habits.

The number of suicides was equal to the number expected. This observation was in line with a previous study in which young adults with congenital heart disease assessed their emotional state as similar to that of the control population (26). In some studies, a severe physical illness has increased suicidal behavior (27,28), but in our study none of the patients with severe heart defect committed suicide. The 2 suicides connected to heart defects should warrant an offer of psychological assistance for patients with difficult health problems.

Conclusions

Increased risk of CHD-related death followed patients throughout their lives. The high number of deaths due to respiratory and neurological causes reflects increased comorbidity. The comorbidity will be studied further among these patients, because the mortality gave just a rough image of it.

The number of accidental deaths was lower among patients with CHD than among the general population. The progress of treatment will likely lower the number of fatal complications among patients in the future.

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